

Successful repair of a popliteal aneurysm with saphenous vein graft in a patient with Marfan syndrome

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ABSTRACT

Marfan syndrome is an autosomal dominant disorder caused by mutations in the fibrillin 1 gene (*FBN1*). This leads to defective elasticity of connective tissue in the arterial wall. Aortic aneurysms and dissections are the most common vascular anomalies; the incidence of peripheral artery aneurysms is not well understood. Treatment options for infrainguinal disease are limited as endovascular interventions are generally contraindicated. The best conduit for arterial reconstruction is also unknown because there is concern that saphenous vein may become aneurysmal. Currently, there are few case reports regarding outcomes of infrainguinal arterial reconstructions, and follow-up has been very short term. We report a rare case of successful repair of a popliteal aneurysm using a saphenous vein graft in a patient with Marfan syndrome. (*J Vasc Surg Cases and Innovative Techniques* 2019;5:393-5.)

Keywords: Marfan; Popliteal aneurysm; Saphenous vein graft

Marfan syndrome (MFS) is an autosomal disorder caused by mutations in the fibrillin 1 gene (*FBN1*), leading to decreased elasticity of the arterial media.¹⁻¹⁵ Thoracic aortic aneurysms and dissections are predominantly seen.^{1,4,5,7,11-13} Because of the systemic nature of the disease, it is reasonable to believe that other arteries are affected. The prevalence of peripheral artery aneurysms (PAAs) in MFS has not been widely studied but is cited to be as high as 67%.⁵

Evidence-based treatment options for peripheral artery disease, in particular infrainguinal disease, are limited. Endovascular interventions are relatively contraindicated because of the risk of damaging an inherently weak artery.^{3,6,8,14} The best conduit for reconstruction is unknown; synthetic grafts have inferior long-term patency rates, and there is concern that vein grafts may become aneurysmal.^{16,17}

We describe an MFS patient with a popliteal artery aneurysm complicated by distal thromboemboli to the foot who underwent bypass using great saphenous vein (GSV). The patient provided permission for publication of his case detail and images.

CASE REPORT

This patient is a 59-year-old man with MFS. He had multiple remote operations, including an aortic composite graft with aortic valve replacement. He later underwent aortobifemoral bypass and concomitant replacement of bilateral common femoral artery aneurysms with prosthetic conduits.

Three years later, he presented with acute-onset left toe pain. Physical examination demonstrated tender, purple pre-gangrenous toes. He had a weakly palpable dorsalis pedis pulse and monophasic Doppler signals in the pedal arteries. Computed tomography angiography of the chest and abdomen with bilateral lower extremity runoff demonstrated a thrombosed 2-cm left popliteal artery aneurysm with tibial artery reconstitution without other disease (Fig).

He was taking warfarin and aspirin at presentation. He was assessed by the authors and believed to be an appropriate candidate for aneurysm exclusion and bypass. He was admitted and maintained on therapeutic anticoagulation and aspirin. Preoperative lower extremity vein mapping demonstrated non-aneurysmal 4.3-mm GSVs bilaterally.

Repair of this aneurysm was performed by a left superficial femoral to below-knee popliteal artery bypass using an ipsilateral, reversed GSV graft. Intraoperatively, 5000 units of heparin were given. The patient's vessels were not friable and handled suture without needing reinforcement. The graft was anastomosed to the superficial femoral artery, tunneled anatomically, and anastomosed to the popliteal artery distal to the aneurysm, both in end-to-side fashion using 6-0 Prolene. The aneurysm was ligated proximally and distally. Pedal pulses were easily palpable once flow was restored.

The patient did not have perioperative complications. His foot pain and pre-gangrenous changes in the toes resolved. He stayed for 7 days postoperatively because of prolonged heparin bridge to warfarin.

He was seen in clinic 2 weeks after discharge, every 3 months with bilateral lower extremity ultrasound for the first 18 months, then every 6 months for a total of 41 months of follow-up. He

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Fig. Coronal view of computed tomography angiography demonstrating a thrombosed left popliteal artery aneurysm.

continues to do well. Clinical and radiographic examinations demonstrate no new disease. The vein graft remains patent without aneurysmal change, as evidenced by serial graft velocities obtained with each study. At 41 months, the velocities proximal, within, and distal to the graft ranged from 34 to 59 cm/s.

DISCUSSION

MFS is an autosomal dominant disease of the connective tissue with multisystem manifestations, in particular the cardiovascular, ocular, and skeletal systems.^{1,2,4,10,11} It is caused by one of more than 1000 mutations identified in the *FN1* gene encoding the fibrillin 1 protein.¹² Prevalence is 1 in 5000, inherited with high penetrance but with variability in its presentation.²

Fibrillin 1 is a structural glycoprotein serving as scaffolding for elastin deposition and formation of elastin fibers.¹ Mutations directly weaken the elastin layer of the artery, particularly in the aorta.^{7,11} Subsequently, the aorta has decreased capacity to tolerate forces created by the heart and is subject to aneurysmal degeneration, dissection, and rupture.^{11,12} The ratio of elastin to stiffer structural proteins like collagen decreases from 60:40 in the thoracic aorta to 30:70 in the periphery.¹⁸ As such, the ascending aorta is the site of first operation in 83.8% of patients at the mean age of 32.4 years.⁴

Thoracic aortic aneurysms lead to devastating sequelae; therefore, early intervention significantly increases life expectancy.^{4,11-13} Despite operative success, patients continue to experience arterial degeneration throughout life and require reoperations.¹⁴ Given the systemic nature of disease and longer life expectancy, it

is reasonable to expect the development of PAAs that may threaten life or limb.

Routine imaging is not generally performed outside of the thoracic aorta; therefore, the true incidence of PAA is unknown. Yetman et al¹⁵ studied 140 MFS patients with computed tomography or magnetic resonance angiography from the skull base to the iliac bifurcation and identified 44 (31%) patients with distal aortic aneurysms or PAAs. Thirteen (29.5%) ultimately sought emergent care. Gaertner et al⁵ used Doppler ultrasound to systematically examine the supra-aortic vessels, upper and lower extremities, and visceral branches for a year to investigate the rate of involvement outside the thoracic aorta. Of 15 patients, 10 (67%) had PAAs; two were at high risk of rupture and required semiurgent repair.

The best reconstructive option for PAAs in MFS patients remains unclear. Endovascular stents are relatively contraindicated because of their theoretically poor stability and limited data available on the exertion of persistent radial forces in an inherently weak artery.^{3,6,14,19} Ince et al⁷ described six patients who underwent aortic stent grafting; only two (33%) had primary success and one died secondary to aortic rupture. Waterman et al¹⁴ described their experience with 19 patients, of whom only six (31.6%) had primary success. Reasons for failure included endoleak, rupture, dissection, and persistent aneurysmal degeneration.

Few case reports exist of MFS patients who have had successful repair of PAAs, but longer follow-up than our experience has not been reported. Latter et al⁹ resected and reconstructed an internal carotid artery aneurysm with an interposition vein graft, without aneurysmal recurrence in 2 years of follow-up. Ohyama et al²⁰ excised an internal carotid artery aneurysm with end-to-end anastomosis with no abnormality 13 months postoperatively. Dolapoglu et al³ bypassed subclavian and axillary artery aneurysms with a 10-mm polyester graft, and Haruki et al⁶ repaired an axillary artery aneurysm with an 8-mm polyester graft. Hatrick et al²¹ successfully bypassed a superficial femoral artery aneurysm with reversed saphenous vein graft but had only 14 days of follow-up. Wolfgarten et al²² described the only other reported popliteal aneurysm in an MFS patient, bypassed with a 6-mm polyester graft with success at 1-year follow-up.

In our patient, we elected not to employ an endovascular technique because of the relative contraindications to its use.^{16,17,23} Given the infrainguinal location, the long-term patency rate of a synthetic graft may have been suboptimal.^{16,17} We instead used the GSV, which has remained patent without aneurysmal degeneration 41 months after surgery. From this experience, GSV appears to be an appropriate bypass conduit in MFS patients and may not experience aneurysmal degeneration seen in arteries with intermediate follow-up.

CONCLUSIONS

MFS is a genetic disorder characterized by fibrillin 1 defect that profoundly affects the vascular system. Currently, no consensus on screening for peripheral artery disease exists; however, as the life expectancy of MFS patients continues to improve, this is an area that should be explored because of the systemic nature of the disease. We believe, in our limited experience, that the GSV may be a feasible, safe conduit for repair of infrainguinal aneurysms in patients with MFS. Further studies looking at long-term outcomes of saphenous vein conduits as arterial bypasses are needed.

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